NEWBORN SCREENING
2004 ANNUAL REPORT
Including Newborn (Blood-spot) Screening
& Newborn Hearing Screening

NEBRASKA HEALTH AND HUMAN SERVICES SYSTEM

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NEWBORN SCREENING FOR INBORN ERRORS OF METABOLISM and INHERITED DISORDERS

The goal of newborn screening for metabolic and inherited disorders is to identify newborns at risk for certain metabolic, endocrine, hematologic and other disorders that would otherwise be undetected until damage has occurred, and for which intervention and/or treatment can improve the outcome for the newborn.

Newborn Screening is a system involving many elements including:

- Education of health care professionals and parents
- Proper and timely collection of quality specimens
- Appropriate and timely transmittal of specimens to the NBS laboratory
- Rapid quality testing methods
- Timely notification of the infants parents
- Timely retrieval of the infant for confirmatory or repeat testing, appropriate referral to specialists family with treatment for these disorders
- Assuring access to needed specialized services and treatment

Each of these components of the system requires ongoing monitoring to ensure quality.

In 2004 newborn screening efforts resulted in successfully identifying and treating 30 newborns affected with disorders in time to prevent problems associated with them:

- 6 babies with partial biotinidase deficiency
- 8 babies with congenital primary hypothyroidism
- 3 babies with duarte variant galactosemia
- 2 babies with hemoglobinopathies (1 Hgb. C disease, and 1 Sickle Hgb. C disease)
- 4 babies with MCAD
- 1 baby with classical PKU & 3 with hyperphenylalaninemia requiring treatment & 2 hyperphenylalanineia requiring only monitoring
- 3 babies with 3-MCC
- 1 baby with tyrosinemia type II

WHAT IS NEWBORN SCREENING?

Newborn screening programs have been around for over four decades in all 50 States and in several countries. The compulsory screening panel varies from state to state but the overall goal is the same which is to prevent or minimize the serious effects of the disorders screened. Depending on the disorder, effects can range from brain and nerve cell damage resulting in severe mental retardation, to damage to the child's liver, spleen, eyes, problems with physical growth, stroke and even death.

In the first few days after a baby is born, 5 drops of blood are collected from a simple heel stick and applied directly to a special filter paper. These blood spots are shipped to the newborn screening laboratory and tested. When a specimen is "presumptive positive" for a disorder, the physician is notified and has the infant come back for a repeat or confirmatory test. Once a diagnosis is made, treatment can begin. Treatment varies depending on the disorder and for some, intervention may be recommended upon learning the initial screening result, prior to obtaining the confirmatory results. Some examples of treatment are: parent education for recognizing signs/symptoms of metabolic crisis, restricting certain foods from the diet, taking a particular vitamin or medication, supplementing a restricted diet with special foods and formula, or preventive antibiotic treatment. Whatever the treatment, the consequences of not beginning treatment in time can be extremely serious for the infant and the family.

The disorders screened are individually rare, so consultation with and/or referral to the appropriate pediatric specialist such as a geneticist, metabolic specialist, hematologist or endocrinologist is always recommended.

Individually each disorder is quite rare. However, collectively as many as 1 in every 800-1000 babies are diagnosed each year in Nebraska and with disorders from the current screening panel!

Nebraska's Newborn Screening System

In 2004, 69 Nebraska birthing hospitals sent specimens to Pediatrix Screening Laboratory under contract with the State of Nebraska to conduct all of the newborn screens. The military base hospital began sending their specimens to Pediatrix as well in December.

Nebraska requires screening for six disorders: Biotinidase deficiency, Congenital Primary Hypothyroidism, Galactosemia, Hemoglobinopathies, MCAD and PKU. All newborns were offered supplemental screening at no extra cost and which required no extra blood. The supplemental screening included results from tandem mass spectrometry screening for about 30 fatty acid, organic acid and amino acid disorders. Educational efforts of physicians and hospital staff using written materials from the Newborn Screening Program helped parents understand their options. Greater than 95% of parents consented to the supplemental screening.

The Newborn Screening Program in the Nebraska Health and Human Services System included, Mike Rooney Administrative Assistant, Krystal Baumert Follow-up Coordinator, and Julie Miller Program Manager. Personnel worked closely with the metabolic specialists Richard Lutz, MD and William Rizzo MD from the University of Nebraska Medical Center for ongoing consultation. In addition quarterly meetings with the Newborn Screening Advisory Committee provided invaluable guidance to the program on several policy and quality assurance issues.

Treatment services received substantial support via the $10 per infant screened fee, State General Funds and Title V Maternal and Child Health Block Grant funds. This included funding for special metabolic formulas, metabolically altered/pharmaceutically manufactured foods, and support for specialty dietitian services and sub-specialist MD consultation services.

National Attention for Newborn Screening in 2004 (potential impact on Nebraska)

Secretary's Advisory Committee & Media Attention

The federal Health and Human Services Secretary Tommy Thompson's "Advisory Committee on Heritable Disorders and Genetic Diseases in Newborns and Children" set the stage for national debate and publicity around newborn screening with meetings discussing the inequities between state newborn screening programs. Also, due in large part to the efforts of parents from several advocacy groups, these disparities were also showcased in the summer on such national programs as NBC's "Today Show" which carried a 4 part series, ABC News and the Montel Williams program. Several newspapers across the country also covered the issues.

ACMG Report

The Health Resources Services Administration's (HRSA) Maternal and Child Health Bureau (MCHB) already had a number of projects underway in an attempt to address some of these disparities. One of the most significant of these was conducted under a cooperative agreement with the American College of Medical Genetics (ACMG). This project developed an evaluation tool, and with worldwide input evaluated over 80 disorders for screening. They subsequently recommended a uniform panel of disorders in a report to the Secretary's Advisory Committee in September, which was accepted by majority vote of the Committee. The report was published electronically in 2005 with requests for public comment.

A perceived goal of this report is to influence the disparity in newborn screening from state to state, and steer screening towards uniformity, so that all newborns benefit equally.

…"Indeed counting has become increasingly problematic to the point that a competition seems to be taking place in which the apparent superiority of a newborn screening program or private laboratory is staked on the sole basis of quantity, with disproportionate consideration given to quality.

This concept has caught the attention of the media that constantly tell the public-at-large that the more conditions that are screened in a particular State, the better the program must be. As a direct consequence of this behavior, the number of conditions is perceived by the public and policy-makers as a scorecard often leading to either inflated or inaccurate figures."

ACMG Report to the Secretary of Health & Human Services Advisory Committee on Heritable Disorders and Genetic Diseases in Newborns and Children

Spring 2005

State Responses

In the meantime States across the nation were rapidly moving forward to add screening by tandem mass spectrometry and other disorders in the recommended uniform panel. Fortunately for Nebraska, all but two of the disorders were already either required, or part of the universally offered supplemental panel.

Nebraska Medical Association Support

In response to the screening panel expansion already experienced in Nebraska and in preparation for the expected ACMG recommendations the Nebraska Medical Association issued a resolution in support of newborn screening. The resolution encouraged the State Health and Human Services System to ensure sufficient resources for the state support services (e.g. follow-up, patient and professional education, treatment) commensurate with the expanded screening. In response the HHSS Policy Cabinet supported a proposal for a bill that would enable the per-infant-screening fee to be adjusted to meet actual costs. This bill however did not get introduced.

Program Moves to Add CF and CAH to the Screening Panel

Following the National March of Dimes announcement about the ACMG recommendations for the uniform panel, the Nebraska Newborn Screening Advisory Committee (NBSAC) began in-depth reviews of the disorders not included in the required or supplemental panels. Work groups on Cystic Fibrosis (CF) and Congenital Adrenal Hyperplasia (CAH) met in November and December and recommended adding these two disorders to the state's required panel. A third work group on the supplemental tandem mass spectrometry disorders also recommended adding the disorders from the ACMG list of uniform disorders to Nebraska's required panel.

MAJOR INITIATIVES of 2004

Education

- Translations of the patient education materials "Parent's Guide to Your Baby's Newborn Screening"and the "Supplemental Consent Form" into Spanish, Vietnamese, Arabic, Chinese and Russian were completed.

- The program continued to provide supplies of the "Parent's Guide" and supplemental newborn screening consent forms to all birthing hospitals and upon request to childbirth educators and clinics.

- Staff from the program worked with a new hospital in Omaha to help prepare their obstetric services for newborn screening.

- Nebraska was represented nationally by program staff presenting at the Association of Public Health Laboratory's National Newborn Screening meeting on the "Public/ Private Partnership" Nebraska's newborn screening system has adopted. This system was also featured in an Association of State and Territorial Health Officers newsletter.

- Educational updates on newborn screening were also presented at these State level meetings: Beatrice State Developmental Center Sharing Conference and the Nebraska Association of Clinical Lab Manager's Spring Conference in Omaha

Policy Development

The Newborn Screening Advisory Committee and State Newborn Screening Program had determined PKU screening could be improved by changing the screening method for PKU from HPLC (High Pressure Liquid Chromatography) to using Tandem Mass Spectrometry. However, because Nebraska previously had a multi-lab system, regulations were specific as to testing methods in order to provide standardization. Now that these decisions could be negotiated via the contract for the laboratory, the need for the prescriptive regulations was not as great. Therefore the Department proceeded with regulation changes to facilitate adoption of improved technologies in screening. These changes were in process when a national emergency hit the newborn screening world. The FDA had taken action against the supplier of the reagent used to screen for PKU with HPLC. More than half the birth population of the U.S. was affected by this action. Fortunately Nebraska had already planned to switch to tandem mass spectrometry testing, and so with an emergency approval of the Governor to advance the proceedings for public hearing, we were able to adopt the changes, and avert disaster before the laboratory ran out of the reagent. In the meantime, the Association of Public Health Laboratories (APHL) worked with the FDA and vendor to resolve the problems.

Quality Assurance

Lab Visit
In February 2004, the program conducted a site visit at the Pediatrix Newborn Screening Laboratory in Pennsylvania. Laboratory expertise was brought to the team by Doug Stickle, Ph.D. from the University of Nebraska Medical Center and medical expertise by Richard Lutz, MD Pediatric geneticist and specialist n metabolism and endocrine disorders. Julie Miller, Program manager and Krystal Baumert, Follow-up Coordinator helped round out the team to look at the operations, administration , data, communications and follow-up aspects of the laboratory. The entire team was impressed by the expertise and resources devoted to quality laboratory testing, information systems and the efforts to meet the needs of Nebraska's newborns through our contract.

Newborn Screening Advisory Committee
The Newborn Screening Advisory Committee (NBSAC) provides technical expertise and policy guidance to the Nebraska Newborn Screening Program. The following summarizes this guidance:

Quality Assurance Reviews:
In 2004, the Committee continued to review quarterly quality assurance reports from the program. The Committee also monitored aggregate data received by the program on supplemental screening using tandem mass spectrometry. Refer to Section III of this report for summaries of this data.

Further Regulation Revision Recommendations:
In the fall of 2004, the Committee responded to the ACMG report recommending certain disorders be included in all State's "uniform" newborn screening panels. Members of the NBSAC were joined by representatives from the Nebraska Medical Association, and Nebraska Hospital Association, on three work groups that conducted in-depth evaluation of Cystic Fibrosis, Congenital Adrenal Hyperplasia, and the additional disorders on the tandem mass spectrometry supplemental screening panel. The CF work group also had representatives from Medicaid, the Certified Cystic Fibrosis Center, a genetic counselor and a parent representative. These work groups prepared summary reports including their recommendations to the Newborn Screening Advisory Committee.

Committee Structure:
The members of the NBSAC in 2004 were/are:

- **Khalid Awad**, MD, *Neonatologist*, Neonatal Care PC, Omaha
- **Lawrence Bausch**, MD, *Neonatologist,* Saint Elizabeth Regional Medical Center, Lincoln
- **Kevin Corley**, MD, *Pediatric Endocrinologist*, Children's Hospital, Munroe/Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Jeanne Egger,** *Parent***,** Hallam,
- **Adolfo Garnica,** MD, *Pediatric Endocrinologist*, Children's Hospital, Omaha
- **David Gnarra,** MD, *Pediatric Hematologist,* Children's Hospital, Omaha

- **James L. Harper**, MD, *Pediatric Hematologist*, UNMC, Omaha
- **Kathryn Heldt,** RD, *Dietitian*, Children's Hospital Metabolic Clinic, Omaha
- **Mary Kisicki,** RN, *Parent*, Papillion
- **VICE CHAIR: Richard Lutz,** MD, *Geneticist, Pediatric Endocrinologist, Pediatric Metabolic Specialist,* Munroe/Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Bev Morton,** *Parent,* Lincoln VICE CHAIR
- **Howard Needleman**, MD, *Neonatologist*, Children's Hospital, Omaha
- **Samuel Pirruccello**, MD, *Pathologist*, Regional Pathology Services, UNMC, Omaha
- **William Rizzo,** MD**,** *Geneticist, Pediatric Metabolic Specialist,* Munroe Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Kathy Rossiter,** MSN, *Certified Pediatric Nurse Practitioner*, Children's Hospital Metabolic Clinic, Omaha
- **G. Bradley Schaefer**, MD, *Geneticist, Pediatric Endocrinologist, Pediatric Metabolic Specialist*, Munroe Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha (ex-officio member)
- **Jill Skrabal,** RD, *Dietitian*, Munroe Meyer Institute for Genetics and Rehabilitation, UNMC, Omaha
- **Douglas Stickle**, PhD, *Technical Director, Clinical Chemistry*, UNMC, Omaha
- **William Swisher,** MD, *Pediatrician,* Lincoln Pediatric Group, Lincoln
- **Thomas Williams**, MD, *Pathologist*, Pathology Center, Omaha
- **B.J.Wilson,** MD, *Neonatologist/Perinatologist*, Saint Elizabeth Regional Medical Center, Lincoln, March of Dimes Representative
- **CHAIR: Hobart Wiltse**, MD, PhD, *Pediatric Metabolic Specialist*, UNMC, Retired, Omaha,

Assurance (access to treatment and services)

Part of the public health assurance role of Newborn Screening is ensuring treatment availability and access. Toward that end, the state program manages several contracts to ensure provision of otherwise prohibitively expensive formulas, foods, and services not always reimbursed by insurers. Fifty nine individuals were served through these programs.

Federal funds allocated to Nebraska under the Title V Maternal and Child Health Block Grant have been used for many years to support nutritional counseling and to provide specialized formulas for individuals with PKU. This has also been extended to special formulas for other metabolic disorders screened for in Nebraska. In addition, State General fund appropriations of $42,000 and funds generated from the screening fee help purchase the specialized formula and pharmaceutically manufactured foods.

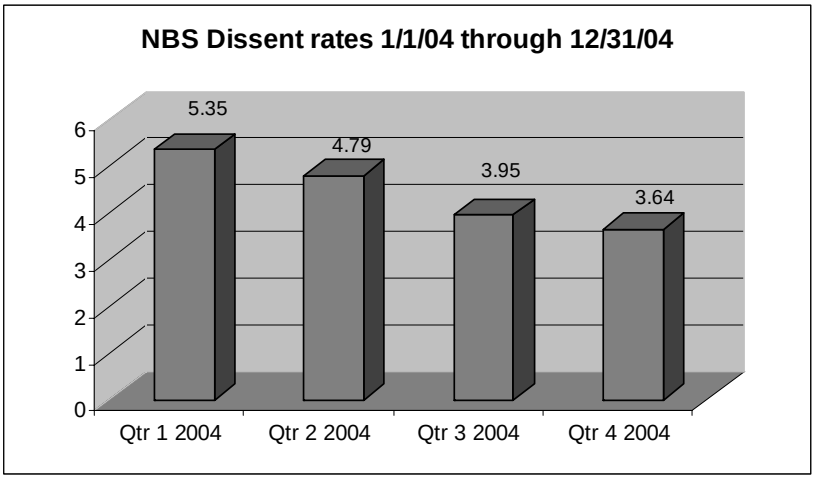
The number of children identified with disorders requiring special formula is anticipated to increase. State General Funds and Maternal and Child Health Title V Block grant funds have been level funded for several years (no increases, and no allowance for inflation). Therefore, the program forwarded a bill proposal that would have enabled adjustments to the $10 per infant screened fee. Since this did not get introduced, the

metabolic formula contract now requires billing the patients insurance (including Medicaid) first, then using these other funds as payer of last resort.

PROCESS/OUTPUT DATA FOR 2004

PATIENT EDUCATION

Consent for supplemental screening



Overall for 2004, 95.57% of parents consented to the supplemental newborn screening panel from MS/MS. Hospital personnel report that since this screen does not require any extra blood, and no additional cost, more parents are requesting it.

Patient education for home births

In 2004, there were 60 home births reported to the Department of Health and Human Services Newborn Screening Program and all of these were screened. Once reported, if the infant has not already been screened, the Department works with the families, physicians, hospitals and laboratories to facilitate getting these infants screened. The state program commits a significant amount of time and personnel resources to educating parents about the mandatory screening law, the benefits of newborn screening and how to get their baby screened.

SPECIMEN COLLECTION, HANDLING AND TRANSPORT

Age at Time of Specimen Collection (Initial Specimen)

Age at time of collection	Number of births	Percent of births
0-12 hours	155	0.59%
12-24 hours	95	0.36%
Collected day 2 (24-48 hours of age)	23,125	88.11%
Day 3	2,371	9.03%
Day 4	227	0.86%
Day 5	40	0.15%
Day 6	22	0.08%
Day 7	21	0.08%
Over 7 days	191	0.73%

Regulations require all specimens to be collected between 24-48 hours of birth, or prior to discharge, transfer or transfusion which ever comes first. Specimens collected past day 2 are at increased risk of a delayed diagnosis.

Unsatisfactory Specimens for 2004

Number of specimens unsatisfactory / Total # initial specimens	111 of 26,443	0.41% of specimens
REASONS specimens were UNSATISFACTORY	**Number**	**% of unsats**
Blood spots not soaked through	26	23.42%
Blood heavily applied, layered or double spotted	23	20.72%
Exposed to heat or humidity	20	18.11%
Serum or fluid mixed with sample	14	12.61%
Specimen contaminated or diluted	11	9.90%
QNS (Quantity not sufficient)	6	5.40%
Clotted	4	3.60%
Surface of blood spots is scratched or abraded	2	1.80%
Blood applied to both sides of paper	1	0.90%
Plasma separated from red blood cells (possible line collection)	1	0.90%
No blood on form	1	0.90%
Specimen got wet prior to arrival at laboratory	1	0.90%
Outdated collection form	1	0.90%

The art and science of correctly collecting and handling dried blood spots on filter paper requires trained health care professionals with strong skills in attention to detail and who consistently follow the Clinical and Laboratory Standards Institute procedures for specimen collection. Every unsatisfactory specimen must be repeated in order to ensure sufficiently reliable screening results.

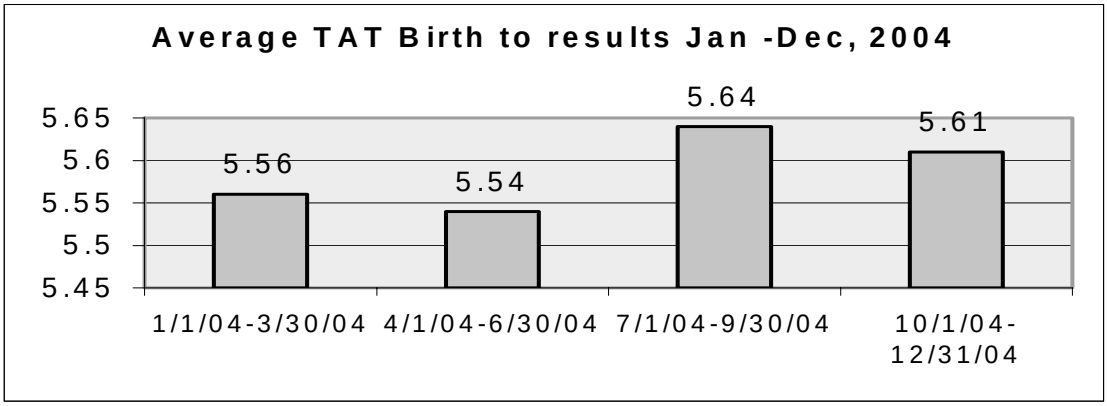
Drawn Early (less than 24 hour) Specimens for 2004

Reason specimen collected at less than 24 hours of age	Number / Percent
Baby to be transferred	90 47%
Baby to be transfused	22 9%
Unable to determine reason from data received at NNSP	117 51%

- Twelve of the drawn early newborns did not get repeated as they expired.
- An additional 62 infants were reported as drawn early but upon notification to the birthing facility it was reported that they had made reporting errors in these cases.

Specimen Turn Around Time

Regular monitoring of turn around time of results reporting from the initial specimen, is an important indicator for how well the newborn screening system is functioning to be able to identify affected infants in time to prevent the effects of the disorder.



LABORATORY TESTING DATA

Presumptive Positive Screening Rates

In 2004 there was some public discussion that misrepresented newborn screening as highly inaccurate. This discussion focused only on the "false positive" rate and failed to recognize that these rates were derived from very small numbers of positive screening results in the first place. It also failed to recognize that screening programs by their very nature are designed to find those at higher risk of a disease in order to facilitate their diagnosis and treatment to prevent morbidity and mortality. Screening tests were never designed to be diagnostic. Despite this fact, programs across the country strive to minimize the number of newborns that require repeat or confirmatory testing (presumptive positive), and maximize the probability of identifying those affected.

Including only the disorders required to be screened (6), times the number of newborns screened (26,391), the number of tests completed for Nebraska newborns were 158,346. Of this 158,346, only 494 were presumptive positive requiring repeat or confirmatory testing. This is an overall presumptive positive rate of only 0.31%. Considering another 95% of newborns also received the supplemental tandem mass spectrometry test for an additional 31+ disorders, (conservatively 25,071 x 31 = 777,201 additional results were available with only 137 of these requiring repeat testing and a handful of those requiring confirmatory testing.

The relative low presumptive positive rates maintained by the Nebraska Newborn Screening Program provides a strong testament to the dedication of the Advisory Committee, Program and Laboratory to providing a high quality newborn screening program.

Specific presumptive positive rates by disorder			
Disorder	**National rate 2001***	**Nebr. 5 year mean average (2000-2004)****	**Nebraska 2004 rates (mean average)*****
Biotinidase deficiency	0.01% 1:10,000	0.03% 3:10,000	0.12% 12:10,000
Congenital Primary Hypothyroidism	1.37% 137:10,000	0.39% 39:10,000	0.23% 23:10,000
Galactosemia	0.11% 11:10,000	0.03% 3:10,000	0.03% 3:10,000
MCAD	.007% <1:10,000	N/A universal screening began 7/03	0.01% 1:10,000
Phenylketonuria	0.07% 7:10,000	0.01% 1:10,000	0.02% 2:10,000

*National Rate 2001 is based on the sum of all reported presumptive positives divided by the sum of all the infants reported screened for the disease specified. This rate is converted from % to X:10,000(rounded) for common reporting purposes. National data source: "2001 National Newborn Screening Report, Initial screening results", Biotinidase, Congenital Hypothyroidism, Galactosemia, MCAD, PKU newborns screened total column and newborns presumed with condition column. For CH, galactosemia and PKU 3 or 4 states reported confirmed cases but not the presumptive positive cases. Caution should be used in comparison of numbers.

**Nebraska's 5-year mean: is the mean of the 5 rates figured for each year individually for 2000 through 2004.

***Nebraska's rate 2004: are the number of presumptive positives divided by the total number of newborns screened each year.

CAVEAT: States use varying instruments, methodologies and cut-offs. In addition, the national data report notes inconsistencies in reporting by some states which brings into question the validity of data. Therefore, direct correlations can not be made from the data that is available. However, from the summary of data on the next page, one can extrapolate that in general, Nebraska's chosen technology, methodologies and cut-offs have resulted in positive screening rates that are reasonable compared to other newborn screening programs across the country. Rates for hemoglobinopathies were not figured

due to variances in reporting methods for the national report, and from states. The national data was published as submitted by individual states, and can be found at the web site for the National Newborn Screening and Genetic Resource Center.

Mean Averages of Laboratory Test Measures

The program continues to provide lab testing data to the Newborn Screening Advisory Committee to monitor ongoing quality. The following graphs depict the quarterly mean averages for biotinidase measures, T_4 the primary screen for Congenital Primary Hypothyroidism, and GALT and total galactose used to screen for Galactosemia. Access to data for mean averages for PKU and MCAD is not yet available from the tandem mass spectrometry results from Pediatrix Screening laboratory. These means can tell us something about stability of the assay, reagents etc. over time.

	Jan-Mar 2004	Apr-Jun 2004	Jul-Sep 2004	Oct-Dec 2004
T_4 mean averages	16.980	15.745	15.784	16.516

	Jan-Mar 2004	Apr-Jun 2004	Jul-Sep 2004	Oct-Dec 2004
Galt mean averages	337.308	282.898	270.831	340.440

	Jan-Mar 2004	Apr-Jun 2004	Jul-Sep 2004	Oct-Dec 2004
Total galactose averages	3.076	2.787	2.599	3.157

	Jan-Mar 2004	Apr-Jun 2004	Jul-Sep 2004	Oct-Dec 2004
Biotinidase mean averages	46.199	41.533	38.376	50.144

NEWBORN SCREENING OUTCOME DATA

	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004
Total Births	23,552	23,471	23,631	23,862	24,209	24,958	25,109	25,515	26,067	26,443
Births Screened	23,533 99.9%	23,455 99.9%	23,627 99.9%	23,858 99.9%	24,118 99.9%	24,863 99.6%	25,043 99.7%	25,478 99.85%	26,008 99.77%	26,391
Total Births Lost to Follow-up	20	16	4	4	9	6 + (89 not screened -as expired @ <48 hours.)*	2 + (64 not screened as expired @ < 48 hours)	5 + (32 not screened as expired @ < 48 hours)	5 + (54 not screened as expired @ < 48 hours)	2 + (50 not screened as expired @ < 48 hours)
Total Births PP	216	356	1,140	547	357	412	432	456	415	499
Home Births	61	78	90	83	86	109	93	99	70	60
Home Births Screened	49	68	86	81	77	105	88	95	65	60
Home Births Lost to follow-up[1]	12	10	4	2	9	4	2 + (3 expired)	2 + (2 expired)	3 + (2 expired)	0

*New match with death records beginning in calendar year 2000, to more accurately report #'s actually screened.

Biotinidase Deficiency	**1995**	**1996**	**1997**	**1998**	**1999**	**2000**	**2001**	**2002**	**2003**	**2004**
Presumptive Positive	32	35	5	3	4	2	4	3	4	34*
Confirmed Negative	31	34	2	2	2	2	1	1	0	29
Confirmed Positive (Profound)	0	1	1	1	1	0	0	2	1	0
Confirmed Positive (Partial no tx)	1	0	0	0	0	0	0	0	0	0
Confirmed Positive (Partial tx)	0	0	2	0	1	0	3	0	3	6

* Screening protocols identified most of these as "inconclusive", for which repeat screening rather than confirmatory testing, ruled out the disorder.

Congenital Primary Hypothyroidism	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004
Presumptive Positive	169	276	771	274	108	114	115	129	89	63
Confirmed Negative	161	262	746	265	92	104	105	113	75	55
Confirmed Positive	8	14	10	6	13	8	7	15	11	8
Confirmatory Lost to follow-up	0	0	15	3	3	2*	3*	1*	3*	0

* Lost to follow-up as baby's expired.

Galactosemia	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004
Presumptive Positive	N/A	9	43	9	13	12	15	5	3	9
Confirmed Negative	N/A (1)	7	29	9	8	8	9	5	0	6
Confirmed Positive (Classical)	1	0	0	0	0	1	0	0	1	0
Confirmed Positive, Duarte (not treated)	N/A	1	6	0	3	1 Duarte Hmzgt	0	0	1	0
Confirmed Positive, Duarte (treated)	N/A	0	6	0	2	2 Duarte Mixed Htrzgt. (1 tx'd 1 year)	6 Duarte Mixed Htrzgt.	0	1	3
Confirmed Neg. Classical/CP carrier	N/A	1	1	0	0	8	0	0	0	0
Confirmatory testing not done[1]	N/A	0	1	0	0	0	0	0	0	0

Hemoglobinopathies	1995	1996	1997	1998	1999	2000	2001	2002	2004	2004
SICKLE CELL DISEASE FS Screened positive	N/A	1	3	1	3	2	4	4	5	0
Confirmed Positive	N/A	1	3	1	3	2	4	4	5	0
SICKLE CELL TRAIT FAS Screened positive	N/A	16	88	54	120	139	146	156	150	171
Confirmed Positive	N/A	16	40	54	60	104	102	111 (+1 other variant)	102	81
Diagnosis Unknown	N/A	N/A	48	0	60	35	44	45	48	90
OTHER CLINICALLY SIGNIFICANT Screened positive	N/A	-	-	1	3	14	21	2	1	4
Confirmed Positive-	N/A	-	-	1	1	2	3	2	1	2
OTHER HEMOGLOBIN VARIANTS Screened positive	N/A	-	-	30	228	106	145	150	153	205

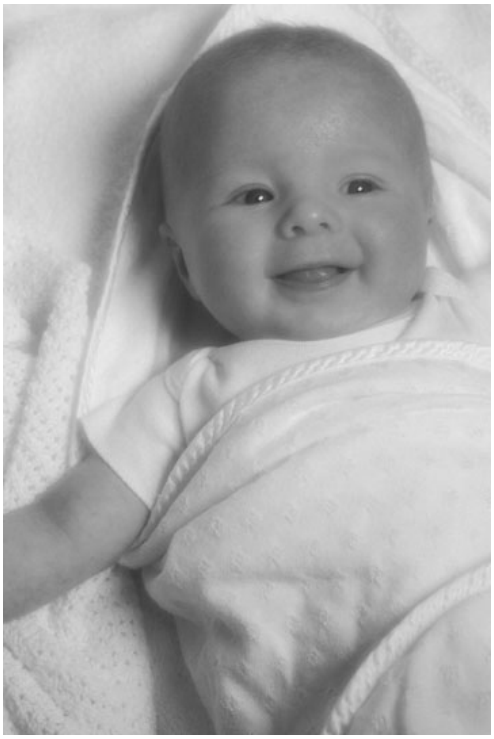
MCAD *	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004
Screened Positive	NA	N/A	N/A	N/A	N/A	N/A	N/A	3*	3	5
Confirmed Negative	N/A	N/A	N/A	N/A	N/A	N/A	N/A	2	3	1
Confirmed Positive	N/A	N/A	N/A	N/A	N/A	N/A	N/A	1	0	4

*Mandatory screening for MCAD began 7/01/2002. Prior to that about 34% of newborns were voluntarily screened in Nebraska in 2000 and 2001, but only numbers the screened were reported to the NNSP.

Phenylketonuria (PKU)	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004
Presumptive Positive	15	14	137	43*	3	6**	4	3	7**	7
Confirmed Negative	14	13	106	40	0	2	2	1	1	1
Confirmed Positive Classical PKU	1	1	3	2	1	1	1	1	2	1
Confirmed Positive Hyperphe			4		2 (tx'd)	1 transient	1	1	3	5 (3 of these tx'd)
Confirmed Positive transient tyrosinemia			24			1	0	0	0	0

1998: One confirmatory testing not done – residence in another state

****2000 and 2003***: One each year for whom confirmatory testing was not done as the baby's expired



Supplemental (Tandem Mass Spectrometry Screening Results)

2004 SUMMARY OF MS/MS FINDINGS (Includes MCAD & PKU, as well as supplemental findings)

Initial findings	**Number Abnormal on screen**	**Number confirmed negative**	**Number confirmed positive**	**Number pending or lost to follow-up**
Tyrosine elevated	37	30	0 (two possible transient tyrosinemia or liver disease) **1 Tyrosinemia type II**	2-lost to follow-up 5-pending repeat or dx.
C3 elevated (propionylcarnitine)	32	31	0	1-pending
C3 & C3:C2, C3:C16 elevated	18	16	0	2-pending
Methionine elevated	13	13	0	0
C5-OH hydroxisovaleryl carnitine	3	0	**3 with 3-MCC** (3-methylcrotonyl Co-A carboxylase deficiency)	0
Generalized elevations of amino acids	8	8	0	0
Phenylalanine or phe + phe:tyr	7	1	**5-hyperphenylalaninemia 1-classical PKU**	0
C8 elevated (octonoylcarnitine)	8	4	**4 with MCAD**	0
C12 elevated (dodecenoylcarnitine)	2	1		1-lost to follow-up
C3:C4 elevated	1	1	0	0
C16	1	1	0	0
Free carnitine and short chain acylcarnitines	1	1	0	0
C14:1 ratio to C16, + other long chain acylcarnitines elevated	1	1	0	0

C16 and C18:1 elevated (palmitoylcarnitine & oleylcarnitine	1	1	0	0
Generalized elevation of short and medium chain acylcarnitines	2	2	0	0
C4 (butyrylcarnitine) elevated	1	1	0	0
Ornithine	1	1	0	0
2004 Totals	**137****	**113**	**13**	**11**

*Lost to follow-up designated when the patient/parent can no longer be found, there is no medical home, or they have moved out of state to an unknown location.
**The vast majority of abnormal screens from MS/MS require only a repeat screen to rule out the disorder. More in-depth confirmatory testing, is recommended in a small percentage of cases where the concentration of analytes are "significantly" abnormal.

Intervention Data

The intervention data is one of the most important measures for determining how well we are doing as a system to ensure timely treatment of affected infants. The following data is grouped by disorder and shows Nebraska's averages/ranges for 2004

The data also includes national averages/ranges according to the most recent available data "National Newborn Screening Report -2000" available at the National Newborn Screening and Genetics Resource Center's web site:
http://genes-r-us.us.uthscsa.edu/resources/newborn/00chapters.html.

Comparisons should be made with extreme caution. States and territories included in the averages in this report, have birth numbers from fewer than 2,000 per year to around 500,000 per year. Likewise, resources necessary to complete testing, follow-up, confirmation, diagnosis and treatment also vary from state to state. The intervention data is one kind of outcome data that can, over time, help to identify how well a state's system is working in newborn screening. The mean average age at time of treatment can be an indicator of whether adequate resources are devoted to each of the components of a comprehensive newborn screening system: education, specimen collection handling and

transportation procedures, laboratory procedures, follow-up and referral procedures, confirmation and treatment.

Biotinidase Deficiency

Nebraska 2004 Intervention data	**U.S. 2000 Intervention data** (most recent national data available)
Goal age for treatment initiation: Upon Diagnosis	24 States/territories screening for biotinidase deficiency
Diagnosed/treated: 6 partials (all partials/treated)	18 cases of biotinidase deficiency reported
Mean Avg. age at Tx. Initiation: 26.16 days	3 or 17% treated by 14 days of age 1 or 6 % treated by 15-21 days of age 6 or 33% treated at > 21 days of age 8 or 44% age of treatment unknown/not reported
Range of ages at Tx. Initiation: 9-45 days	Range of ages @ tx.: 9 - > 21 days

From Nat'l NBS report, table 8.06

Congenital Primary Hypothyroidism

Nebraska 2004 Intervention data	**U.S. 2000 Intervention data** (most recent national data available)
Goal age for treatment initiation: As early as possible, upon diagnosis.	53 States/territories screening for Primary Hypothyroidism
# diagnosed/treated: 8	1,663 Cases of Primary Hypothyroidism detected
Mean Average Age at Treatment initiation: 18.25 days age	Age at treatment: 562 or 34% treated by 12 days of age 264 or 16% treated between 13-21 days of age 298 or 18% treated at > 21 days 539 or 32% age at tx. Unknown or not reported
Range of ages at Treatment initiation: 6-20 days of age	Range of ages at treatment initiation <3 to > 21

From National NBS report, Table 4.12

Galactosemia

Nebraska 2004 Intervention data	**U.S. 2000 Intervention data** (most recent national data available)
Goal age for treatment initiation: As early as possible, upon diagnosis. Diet intervention upon positive screening result. # diagnosed/treated classical and 0 Duarte/mixed heterozygote 3 Mean avg. Age at treatment Initiation: Intervention with soy formula: 9.33 Seen by specialist: 9.66 Range of ages at Tx. Initiation: 6 – 15	52 States/Territories screening for Galactosemia 60 cases of classical galactosemia identified Age at treatment: 17 cases or 28% treated at: 4 days or less 50 cases or 83% treated by: 21 days of age 2 cases or 3% treated at: > 21 days of age 8 cases or 13% age at tx: unknown or not reported Range of ages at treatment initiation: < 3 days - > 21 (Age at treatment for galactosemia variants not reported nationally.)

From National NBS report, Table 5.08

MCAD - Medium Chain Acyl Co-A Dehydrogenase Deficiency

Nebraska 2004 Intervention data	**U.S. 2000 Intervention Data**
Goal age for treatment / intervention initiation: As early as possible, upon positive screening result – parent education/ consultation. # diagnosed/treated: 4 Average age at intervention: 4 days Range in age at intervention: 3-6 days	No data reported for MCAD Deficiency

PKU - Phenylketonuria (Classical PKU)

Nebraska 2004 Intervention data	**U.S. 2000 Intervention data** (most recent national data available)
Goal age for treatment initiation: As soon as possible but no later than 7-10 Days after birth.* # classical PKU: 1 Hyperphe: 3 Avg. Age at treatment classical: 15 Range ages at treatment: 15 Avg. Age at treatment hyperphe: 11.66 Range ages at treatment: 5-18 days 25% treated by 7 days of age.	Cases of classical phenylketonuria 189 55 or 29% treated by 7 days of age 68 or 36% treated between 8-14 days of age 26 or 14% treated between 15-21 days of age 13 or 7% treatment at > 21 days of age 27 or 14% age at treatment unknown or not reported

*NIH Concensus Statement October/25/2000: Phenylketonuria: Screening and Management/ Nat'l data 3.10

Hemoglobinopathies:

Nebraska 2004 Intervention data	**U.S. 2000 Intervention data**[1]
Goal age for treatment initiation:[2] 60 days of age or less # Cases diagnosed/treated SC disease 1 Hgb. C disease 1 Mean/Average age (days) at treatment: SC 20 C diseae 32 Range of ages (days) at treatment: 20-32 100% treated by 60 days of age.	Cases of sickle cell disease confirmed (FS): 885 # cases diagnosed by 60 days of age: 485 or 55% # of cases for which age at treatment was unknown or not reported: 315 or 36% - Cases of sickle hemoglobin C disease (SC) confirmed: 442 # of cases diagnosed by 60 days of age: 304 or 69% # of cases for which age at diagnosis was unknown or not reported: 33 or 7.5%
NOTE: Age at treatment is measure for Nebraska data.	NOTE: National data does not report age at treatment, but reports age at DIAGNOSIS.

[1]National intervention data reported in these day ranges 0-15, 16-30, 31-45, 46-60, 61-75, 76-90 >90. From tables 13.02 and 13.04

[2] Treatment guideline from A Clinical Practice Guideline #6, Sickle Cell Disease: Screening, Diagnosis, Management and Counseling in Newborns and Infants, U.S. Dept. Of Health and Human Services, Public Health Service, Agency for Health Care Policy and Research.

PLANS

Screening Panel Expansion: Nebraska now screens nearly 100% of newborns for six disorders and about 95% of newborns for an additional 31 plus organic acid, fatty acid and amino acid disorders that can be detected on tandem mass spectrometry screening. (Counting sub-classes of disorders this number approaches near fifty disorders). The State Program will carry forth the recommendations of the Advisory Committee to add Cystic Fibrosis and Congenital Adrenal Hyperplasia to the required newborn screening panel. This will make Nebraska's NBS program consistent with the recommendations of the American College of Medical Genetics report of screening for (or universally offering screening for) all disorders in their recommended "uniform" panel and "secondary" panel of disorders.

Extensive planning and collaboration will be undertaken in 2005 to ensure a public process for the regulations, adequate patient and professional education, and preparation for birthing facilities, development of infrastructure for access to confirmatory testing and specialty treatment services for the new disorders.

CONTINUING ACTIVITIES

Education: Educational activities from the NNSP will continue through publication of the Annual Report, and as needed through hospital and physician mailings. Opportunities for on-site education are always available upon request from hospitals. Recommendations for improving the Newborn Screening patient education materials will be sought.

Laboratory Testing: The contract with Pediatrix Screening laboratory is a one year contract, renewable for five years. Annual renewals are dependent on the Department's assessment of contractor's performance.

Follow-up/Tracking and Referral: Procedures used by the NNSP to track every newborn to be sure they received an appropriate screen, to follow-up on all transferred, drawn early, transfused, unsatisfactory, and presumptive positive specimens, facilitate confirmatory testing and referral for diagnostic and treatment services will continue. The duties of the Program's Administrative Assistant have changed to include more assistance to the follow-up coordinator since the expansion of NBS into the tandem mass spectrometry arena. The reassignment of duties of a Title V MCH Block grant funded position to newborn screening will assure adequate resources for follow-up with the addition of cystic fibrosis and congenital adrenal hyperplasia.

Confirmatory Testing: The program will continue to work with specialists and the Newborn Screening Advisory Committee to ensure procedures recommended for confirmatory testing are communicated effectively to practitioners. As the screening panel expands, new information will be developed and shared to facilitate practitioner's

understanding of "next steps" when a newborn requires further testing to determine the metabolic condition. Recommended protocols for communicating "next steps" to physicians with newborns who have positive or inconclusive results for CF or CAH will be clearly developed and in place by the time screening begins.

Diagnosis: Practitioner's are strongly urged to consult with the pediatric specialist appropriate to the disorder for which a newborn has a positive screening result. The program will help link the newborn's primary care provider with specialists when needed.

Treatment: Access to treatment was an issue the program had requested the Newborn Screening and Genetics Planning Grant Advisory Committee to review. Funding sources exist for pharmaceutically manufactured foods and formulas for patients with PKU. However, there are some known gaps, e.g. funding for sickle cell and sickle cell trait genetic services, payment sources for routine blood phenylalanine levels for children and women of childbearing age. There are other areas for which we don't know the issues with accessing treatment, e.g. affordability/insurance coverage of levothyroxin for patients with congenital primary hypothyroidism. The Program will continue to monitor the issues associated with access to treatment and seek ways to ensure funding is sufficient to meet affected individual's needs.

Quality Assurance Monitoring: The Program and Advisory Committee will continue to review and act on quarterly quality assurance plan data as well as respond to trends identified with any problems in the interim periods. The laboratory has agreed to develop a QA report at the request of the program, that can be sent to individual hospitals for their own evaluation and comparison with statewide numbers. The program plans to provide these reports beginning in 2005.

NEWBORN HEARING SCREENING

Why Is This Report Important?

Significant hearing loss is the most common birth defect with an estimated incidence rate of one to three per thousand live births. Left undetected, hearing loss in infants can negatively impact speech and language acquisition, academic achievement, and social and emotional development. Before newborn hearing screening, many hearing losses were not diagnosed until 2 ½ to 3 years of age. If detected early, however, the negative impacts can be diminished, and even eliminated, through early intervention. Recent studies have consistently shown that children who were identified with a hearing loss later in childhood have delays in the development of speech, language, social and academic skills compared with those identified during the first six months of age.

Newborn hearing screening is an essential preventative public health program. It meets the following prerequisites for a population screening program –

- Condition is sufficiently frequent in the screened population
- Condition is serious or fatal without intervention
- Condition must be treatable or preventable
- Effective follow-up program is possible

In 2000, the Infant Hearing Act established newborn hearing screening in Nebraska. The statute requires hospitals to educate parents about newborn hearing screening, encouraged hospitals to voluntarily begin screening newborns for hearing loss, and, by December, 2003, to include hearing screening as part of its standard of care and to establish a mechanism for compliance review. The Act also requires that regulations be promulgated to mandate newborn hearing screening if, by December 2003, less than 95% of newborns in the state were receiving a hearing screening. This report presents the status of newborn hearing screening in Nebraska during 2004 (see Part II, Nebraska Newborn Hearing Screening Data for 2004).

What Is Newborn Hearing Screening?

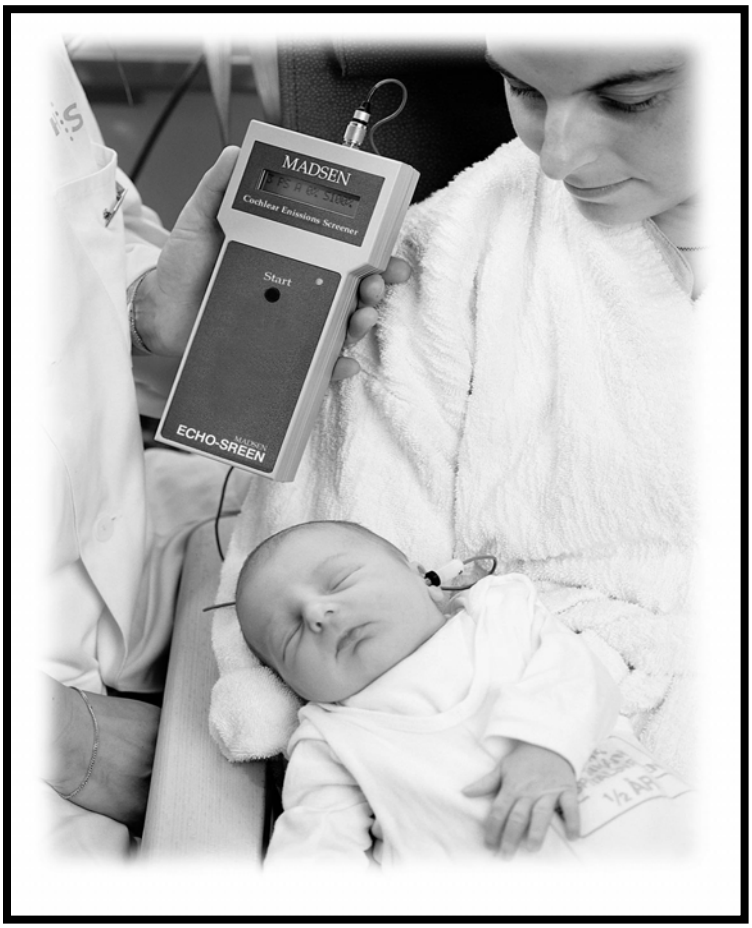
Newborn hearing screening requires objective physiologic measures to detect hearing loss in newborns and young infants. There are two basic measures that birthing hospitals in Nebraska use to screen newborns for hearing loss. Both are easily recorded in newborns and are noninvasive measures of physiologic activity that underlie normal auditory functioning.

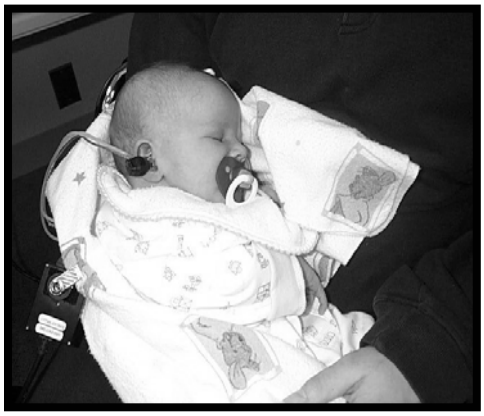
The most frequently used screening technique is measurement of otoacoustic emissions, or OAEs. A miniature earphone and microphone are placed in the newborn's ear canal, low intensity sounds are presented, and responses produced by the inner ear are measured. The second screening technique, Auditory Brainstem Response, or ABR, uses band aid®-like electrodes to detect certain brainwaves in response to sounds that are presented by a miniature earphone. For both methods, the response of each ear is

measured. OAE and ABR are both reliable and accurate. Screening can occur as early as 12 hours of age, preferably with the newborn sleeping, and averages from five to 20 minutes to complete.

If a response is not detected for one or both ears, the result is a "refer" (did not pass). A "refer" to the screening test indicates that a hearing loss *may* exist but there are also other factors that may have contributed. A "refer" does indicate that a second screening is necessary to determine if the other factors, such as vernix in the ear canal, fluid in the middle ear cavity, movement, equipment failures, or inexperience of the tester, contributed to the initial result. A "refer" on the second screening usually indicates the need for a diagnostic audiological evaluation to confirm or rule out a hearing loss and, if hearing loss is present, to begin to identify the type and degree of the loss.

Each birthing hospital has established a newborn hearing screening protocol that identifies how the screening will be administered, the recording and reporting procedures, how refers will be handled, i.e., re-screen as an inpatient with the same or different screening technique or re-screen as an outpatient, and quality assurance measures.





THE NEWBORN HEARING SCREENING SYSTEM

System Elements

The newborn hearing screening system in Nebraska is composed of five functional elements working together to fulfill the purposes of the Infant Hearing Act (Neb. Rev. Stat. §71-4735):

- "To provide early detection of hearing loss in newborns at the birthing facility, or as soon after birth as possible for those children born outside of a birthing facility,
- to enable these children and their families and other caregivers to obtain needed multidisciplinary evaluation, treatment, and intervention services at the earliest opportunity, and
- to prevent or mitigate the developmental delays and academic failures associated with late detection of hearing loss; and
- to provide the state with the information necessary to effectively plan, establish, and evaluate a comprehensive system for the identification of newborns and infants who have a hearing loss."

Newborn hearing screening is one aspect of a comprehensive, integrated Early Hearing Detection and Intervention (EHDI) system. The first three principles of the Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (Joint Committee on Infant Hearing, 2000) are:

1. All infants have access to hearing screening using a physiologic measure. Newborns who receive routine care have access to hearing screening during their hospital birth admission. Newborns in alternative birthing facilities, including home births, have access to and are referred for screening before 1 month of age. All newborns or infants who require neonatal intensive care receive hearing screening before discharge from the hospital. These components constitute universal newborn hearing screening (UNHS).
2. All infants who do not pass the birth admission screen and any subsequent rescreening begin appropriate audiologic and medical evaluations to confirm the presence of hearing loss before 3 months of age.
3. All infants with confirmed permanent hearing loss receive services before 6 months of age in interdisciplinary intervention programs that recognize and build on strengths, informed choice, traditions, and cultural beliefs of the family.

   These three major principles serve as the foundation for the screening, referral, and audiological evaluation protocols developed by the Nebraska Newborn Hearing Screening Advisory Committee in 2001. The timelines established by the NNHSP Advisory Committee are for hearing screening to be completed by 1 month of age, audiological diagnostic evaluation to begin prior to six weeks of age and be completed prior to three months of age, and appropriate early

intervention activities to be initiated by six months of age. The logic model of the NNHSP on the preceding page describes the resources and activities needed to produce the projected results of the program.

The five functional elements of the Nebraska Early Hearing Detection and Intervention system are: Hearing Screening at Birth, Confirmatory Testing, Medical Evaluation, Early Intervention, and Tracking and Surveillance. One or more groups of professionals in a variety of settings assume responsibility of each element of the system. An overview of each of the elements and the primary activities are presented below. Included in this discussion are the Nebraska Revised Statute citations and the recommended protocols established by the Department of Health and Human Services through the Nebraska Newborn Hearing Screening Advisory Committee.

Hearing Screening at Birth

Birthing hospitals in Nebraska have five primary activities related to screening the hearing of newborns:

1. The parent(s)of newborns are educated about the hearing screening, the likelihood of hearing loss in newborns, the importance of follow-up, community resources (including early intervention services), and normal auditory, speech and language development (Neb. Rev. Stat. §71-4740). If risk factors are present, hospital staff educate parents to evaluate hearing every six months. *Note:* The Department of Health and Human Services is responsible for educating the parent(s) for newborns not born in a birthing facility (Neb. Rev. Stat. §71-4740).
2. A hearing screening test is part of each birthing hospital's standard of care for newborn effective 12/1/03 (Neb. Rev. Stat. §71-4742). Following hospital protocol for the procedure, each newborn's hearing in both ears is screened during birth admission using OAE and/or ABR screening techniques. A second inpatient screening is conducted within one to three weeks if the baby "refers" on the first screening. The outpatient re-screening for those that "refer" during birth admission may occur at the birthing facility or at a confirmatory testing facility.
3. A mechanism for compliance review is established for each birthing facility (Neb. Rev. Stat. §71-4742).
4. Results of the hearing screening for each newborn are reported to the newborn's Primary Care Provider. Weekly tracking reports are submitted to the NNHSP that identify newborns who "refer," transfer, or discharge without a hearing screening.
5. Annual reports are submitted to the NNHSP that indicate the following numbers: born in the birthing facility, recommended for screening, received screening during birth admission, passed screening, did not pass screening, and recommended for monitoring and follow-up (Neb. Rev. Stat. §71-4739).

Confirmatory Testing

Newborns who have referred for one or both ears on the second hearing screening should receive an audiological diagnostic evaluation prior to reaching three months of age. The purpose of this evaluation is to confirm the presence of a hearing loss and to determine the type and degree of the hearing loss. The primary activities that comprise the confirmatory testing component are:

1. An initial diagnostic evaluation using either OAE or ABR conducted as early as possible after referral, preferably before the infant is six weeks old. If the infant "passes" this initial part of the evaluation (outpatient re-screening), no further evaluation is usually needed.
2. If the infant "refers" on the initial part of the evaluation, the testing often proceeds immediately to a comprehensive diagnostic evaluation. This evaluation minimally includes measures of middle ear function (tympanometry), auditory sensitivity (air- and bone-conducted ABR), confirmatory measures (parent observations), and, depending upon the developmental age, behavioral audiological assessment (Visual Reinforcement Audiometry). Other measures may be included, as indicated.
3. Depending upon a variety of factors, referrals are made for further evaluation, diagnosis, treatment, and services. These referrals may be made to medical specialists and/or Early Intervention Services.
4. Results of the initial and comprehensive audiological diagnostic evaluation are provided to the Primary Care Physician and NNHSP.
5. Annual reports are submitted to the NNHSP that indicate the number of newborns: who return for follow-up testing, who do not have a hearing loss and who do have a hearing loss (Neb. Rev. Stat. §71-4739).

Medical Evaluation

The infant's Primary Care Provider (PCP) has the key role in the follow-up for those who "refer" on the initial hearing screening during the birth admission. Building on the concept of a pediatric medical home (Guidelines for Pediatric Medical Home Providers, AAP), the PCP has the primary role in identifying and accessing all the medical and non- medical services needed to help children and their families achieve their maximum potential. The primary activities that comprise the medical element of the newborn hearing screening system are:

1. Birthing hospital notifies PCP of the newborn's hearing screening results.
2. NNHSP notifies PCP about the hearing screening status and need for follow-up evaluation for those that did not pass the inpatient hearing screening or were discharged without a screening.
3. PCP or designee per hospital procedure informs parents of hearing screening results and need for re-screening.
4. PCP (or staff), hospital, or test provider schedules re-screen appointment to be completed in one to three weeks and notifies parents.

5. Provider of outpatient re-screening notifies PCP of results.
6. PCP notifies NNHSP of outpatient hearing re-screening results.
7. If "refer," PCP makes referral for comprehensive diagnostic evaluation, educates parents about need for evaluation, and makes referral to Early Intervention services.
8. If hearing loss is confirmed, PCP or diagnostic evaluator refers newborn/infant for complete medical and/or neuro-sensory evaluation and Early Intervention Services.

Early Intervention

Early Intervention is an individualized program of services and supports based on the needs of the individual and family. Part C of the Individuals with Disabilities Education Act (IDEA) authorizes the creation of early intervention programs for infants and toddlers with disabilities. In Nebraska, the Early Development Network (EDN) provides service coordination for eligible families to identify and link with needed services, to work with multiple providers to ensure that services are provided, and to become coordinators of services in the future. The recommended protocols for the primary Early Intervention activities within the newborn hearing screening system are:

1. PCP or diagnostic evaluator makes referral to Early Development Network (EDN).
2. EDN reviews for eligibility.
3. If eligible, EDN may provide assistance with diagnostic evaluation and treatment.
4. Services Coordinator may facilitate obtaining services from otologists, audiologists, community services, and others.

Tracking and Surveillance

The Nebraska Newborn Hearing Screening Program has been developed based on the requirements identified in the Infant Hearing Act (Neb. Rev. Stat. §71-4735 - §71-4744) and the NNHSP Advisory Committee recommended protocols to "…determine and implement the most appropriate system…to track newborns and infants identified with a hearing loss" and "…to effectively plan and establish a comprehensive system of developmentally appropriate services for newborns and infants who have a potential hearing loss or who have been found to have a hearing loss and shall reduce the likelihood of associated disabling conditions" (Neb. Rev. Stat. §71-4737). Activities of the NNHSP include:

1. Develop, implement, and monitor statewide systems to track newborns with or at-risk of hearing loss (Neb. Rev. Stat. §71-4737) and adopt and promulgate rules and regulations to implement the Infant Hearing Act (Neb. Rev. Stat. §71-4742 and §71-4744).
2. Gather required data and generate annual reports (Neb. Rev. Stat. §71-4739 and §71-4741).

3. Establish guidelines for referral to early intervention services (Neb. Rev. Stat. §71-4743).
4. Educate parents with out-of-hospital births about newborn hearing screening (Neb. Rev. Stat. §71-4740)
5. Apply for all available federal funding to implement the Infant Hearing Act (Neb. Rev. Stat. §71-4738).

NEWBORN HEARING SCREENING DATA FOR 2004

Birthing Facilities Data for 2004

Birthing Facility Screening Programs

The number of birthing facilities conducting newborn hearing screening has increased rapidly since 2000 when only 11 hospitals were conducting either targeted or universal newborn hearing screening. In 2004, 100% of the birthing facilities in Nebraska were conducting hearing screenings, consistent with the Neb. Rev. Stat. §71-4742 requirement that a hearing screening test be included as part of the standard of care for newborns. Sixty five of the birthing hospitals were conducting the hearing screening during the birth admission and two were conducting the screening on an outpatient basis following discharge. In 2002 and 2003, sub-grants of $2000 each were provided through the Nebraska Health Care Cash Fund to 38 hospitals with less than 500 births annually to purchase hearing screening equipment.

Birthing Facilities Conducting Newborn Hearing Screenings (2000-2004)

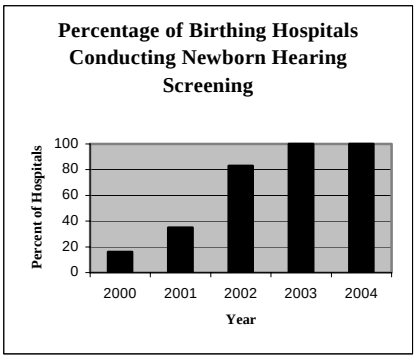


Chart 1

Year	Number of Birthing Facilities in Nebraska	Number Conducting Newborn Hearing Screening	Percentage Conducting Newborn Hearing Screening
2000	69	11	16%
2001	69	24	35%
2002	69	57	83%
2003	67	67	100%
2004	67	67	100%

Table 2

As discussed previously in this report, there are two measurement techniques used to conduct newborn hearing screening: Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR). Half of the birthing hospitals in Nebraska are using OAE-only, almost one third are using ABR-only, and the remaining birthing hospitals are using a 2-step method (OAE, followed by ABR if the initial screening is a "refer"). As can be seen in Table 2, the "refer" rates differ for the three approaches, with the OAE-only having the highest refer rate. The combined refer rate for all of the hospitals is 3.5%.

Refer Rates for Hearing Screening Techniques (2004)

	OAE-only	ABR-only	2-Step
Number of Birthing Facilities	32	25	10
Number of Newborns Screened	3,798	9,938	12,601
Number of Newborns "Referred"	386	309	223
Refer Rate	10.2%	3.1%	1.8%

Table 2

ANNUAL BIRTHING FACILITY REPORTS

Birthing facilities are required to annually report specific information about their newborn hearing screening programs to the Department of Health and Human Services (Neb. Rev. Stat. §71-4739). Reports with aggregate numbers were received from all birthing facilities in 2004.

Birthing Facility Reports of Required Data

	2004
Number of newborns born	26,485
Number of newborns and infants recommended for a hearing screening test	26,447
Number of newborns who received a hearing screening test during birth admission	25,966
Number of newborns who passed a hearing screening test during birth admission, if administered	24,967
Number of newborns who did not pass a hearing screening test during birth admission, if administered	918
Number of newborns recommended for monitoring, intervention, and follow-up care	793

Table 3

The data in Table 3 are based on annual aggregate data reported by the birthing hospitals. Individual screening results and demographic data are not reported for all births. The NNHSP only receives specific information about newborns that "refer" on the initial hearing screening and about those that were discharged without receiving a hearing screening during the birth admission. The opportunity for error exists within the current manual tracking system due to reporting errors, recording errors, duplicated entries because of newborn name changes, transfers from birth hospitals to NICUs, and whether newborns who expire are included in the weekly and/or annual reports. Without a system to accurately determine the status of each newborn's hearing screening results, errors will be present in spite of the best efforts of everyone involved to provide accurate information.

Parent Education

Recommending a hearing screening test has been operationally defined as educating parents about newborn hearing screening, as required by Neb. Rev. Stat. §71-4740. The NNHSP provides print and video education materials free of charge to hospitals to help fulfill this requirement. During 2001 and 2002, the birthing hospitals reported that 92% of parents were educated about hearing screening. In 2003, as the number of hospitals offering hearing screening increased to 100% by the end of the year, the number of hearing screenings recommended through parent education increased to 96.7%. Almost all parents (26,447 or 99.8%) were educated about newborn hearing screening in 2004.

Newborns Receiving a Hearing Screening

The Infant Hearing Act requires that rules and regulations be adopted and promulgated if at least 95% of the newborns in Nebraska do not have a hearing screening by December 1, 2003, or at any time thereafter. The annual aggregate reports submitted by the hospitals in 2004 show that 98.2% of the 26,443 births registered with Vital Statistics were screened during birth admission. The numbers of newborns screened during birth admission has increased dramatically since reporting began in 2000, when only slightly more than one third of newborns received a hearing screening during birth admission (see Chart 1 and Table 4). This increase in the numbers of newborns receiving a hearing screening corresponds to the increase in the number of hospitals adopting newborn hearing screening as the standard of care for newborns and the support of sub-grants through the Nebraska Health Care Cash Fund to purchase screening equipment.

Newborns Receiving a Hearing Screening

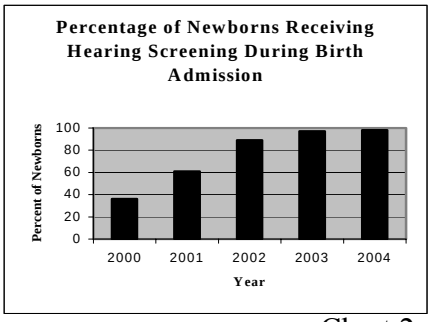


Chart 2

Number and Percentages of Newborns Receiving Hearing Screening

2000	2001	2002	2003	2004
8,978	15,272	22,615	25,275	25,966
36%	61%	89%	97%	98%

Table 4

Newborns Discharged Without a Hearing Screening

During 2004, the annual aggregate hospital reports to NNHSP indicated that there were 188 newborns who did not receive a hearing screening during birth admission because of invalid results (technical or equipment problems), could not test (baby too active/restless), or discharged before screening. Of those 188, there were 158 newborns who were reported to NNHSP on weekly reports as having been discharged prior to screening and 85.4% (135) of them received an outpatient hearing screening at a later time. Of the newborns who received the outpatient screening, the average time to the hearing screening was 23.8 days with 104 (77%) of the newborns receiving the initial screening prior to one month of age.

There were 5 parents who refused the inpatient newborn hearing screening and 162 newborns who expired prior to being screened, according to the aggregate reports. Birthing facilities reported that 664 newborns were transferred to other hospitals (within Nebraska and to surrounding states) prior to hearing screening. Hospitals with Neonatal Intensive Care Units reported receiving 741 transfers from birthing facilities within Nebraska and surrounding states. Including transfers to the NICUs within the birthing facility, 870 transferred newborns were tracked and eight of those were later identified with a significant permanent hearing loss.

Out-of-Hospital Births

Neb. Rev. Stat. §71-4740 requires the Department of Health and Human Services to educate parents of newborns who are not born in a birthing facility about the importance of newborn hearing screening and to provide information to assist them in having the screening performed within three months after the child's birth. Although parent education was provided to the parents of all reported out-of-hospital births during 2004, less than 22% (13) of the 60 out-of-hospital births were screened (see Table 5 for 2001-2004 results).

Out-of-Hospital Births

	2001	2002	2003	2004
Out-of-hospital births	93	80	68	60
Number screened	5	11	10	13
Percentage screened	5.4%	13.8 %	14.7 %	21.7 %

Table 5

Birth Admission Refer Rates

The annual aggregate reports received from the birthing facilities indicated that 918 newborns did not pass (refer) the hearing screening during birth admission. The actual NNHSP file count of newborns who referred was 901. These were the newborns who were tracked through follow-up re-screening and diagnosis, if necessary.

Of the newborns with hearing screenings conducted during the birth admission, the refer rate was 3.7% during 2002, 3.6% during 2003, and 3.5% in 2004. These overall refer rates compare favorably with national statistics that indicate a refer rate of 3.4% for the 36 states with refer rates of less than 5% during the latter half of 2003 (NCHAM 2004 State EHDI Survey).

Monitoring, Intervention, and Follow-up

The final data reported by the birthing hospitals is the number of newborns recommended for monitoring, intervention, and follow-up care: 709 (85% of refers) in 2002, 676 (74% of refers) in 2003 and 793 (86% of refers) in 2004. The reporting of the data for this item seems to be inconsistent with approximately 75% of the hospitals recommending that newborns with a "refer" status on the initial hearing screening need additional monitoring, intervention, and follow-up. The remaining hospitals report very few recommendations for monitoring, intervention, and follow-up care. Beyond the lack of a clear operational definition for this item, the reasons for fewer recommendations than "refers" is unknown.

AUDIOLOGICAL/CONFIRMATORY TEST PROVIDER DATA FOR 2004

Neb. Rev. Stat. §71-4739 requires confirmatory testing facilities to report the following information:

- Number of newborns and infants who return for a followup hearing test
- Number of newborns and infants who do not have a hearing loss based upon the follow-up hearing test
- Newborns and infants who are shown to have a hearing loss based upon the follow-up hearing test

The Advisory Committee for the NNHSP, consistent with recommendations of the American Academy of Pediatrics and the National Center for Hearing Assessment and Management (Guidelines for Pediatric Medical Home Providers, AAP), identified the initial level of the follow-up hearing test as an outpatient re-screening of the newborn's hearing. For those newborns and infants who pass this initial level of the follow-up hearing test, no further audiological evaluation would be needed, unless there are risk factors present that would warrant periodic evaluation. The Advisory Committee recommends that the re-screening occur within the first six weeks to minimize the need to sedate the infant to obtain reliable results and so that intervention can begin early if a hearing loss is identified.

Since the majority of newborns will pass this second screening, considerable cost savings can result by using either the OAE and/or ABR screening technique rather than proceeding directly to a complete diagnostic audiological evaluation. The Advisory Committee's Audiological Diagnostic Protocol recommends that the referral center should be prepared to provide comprehensive audiological diagnostic procedures if the outpatient re-screening results indicate a "refer" status. However, some communities that do not have audiology services readily available have opted to have the initial re-screening occur at the birthing hospital on an outpatient basis.

Newborn Hearing Screening Annual Confirmatory Testing Facility Reports

Each year data regarding the follow-up hearing tests at confirmatory testing facilities have been gathered by surveying the audiologists in Nebraska. Seventeen confirmatory testing facilities responded, representing 44 licensed audiologists. The results of those surveys for 2004 are included in Table 6.

Required Follow-up Hearing Test Data Reported by Audiologists Table 6

Number of audiologists responding	44	
	Re-screenings	Diagnostic Evaluations
Number of newborns/infants receiving a follow-up hearing test	506	110
Number of newborns/infants without a hearing loss	417	53
Number of newborns/infants with a hearing loss	83 (refer)	51

Rate of Follow-up Outpatient Screening and Confirmatory Testing

As an increasing number of birthing hospitals in areas of the state without audiology services nearby are conducting the initial outpatient re-screenings, those figures are important to present a comprehensive view of the follow-up services being provided in Nebraska. In aggregate reports, the birthing facilities indicated that 594 newborns had received outpatient hearing screenings and confirmatory testing facilities indicated that 506 newborns received screenings for a total of 1,100 outpatient screenings. With aggregate reporting, it is not possible to determine an unduplicated count, since some infants, especially those with middle ear dysfunction and an accompanying temporary conductive hearing loss, may be screened several times at one or more sites.

Unduplicated file counts based on patient-specific reports submitted by birthing facilities and confirmatory testing facilities to NNHSP indicate that follow-up screening and/or diagnostic evaluations were initiated for 940 of the 1060 newborns who either did not pass or were discharged without a hearing screening during birth admission. The remaining newborn files are open with follow-up activities either not initiated or reported to NNHSP, lost to follow-up in which the family has moved or no valid contact information is available, or parents have refused additional follow-up activities.

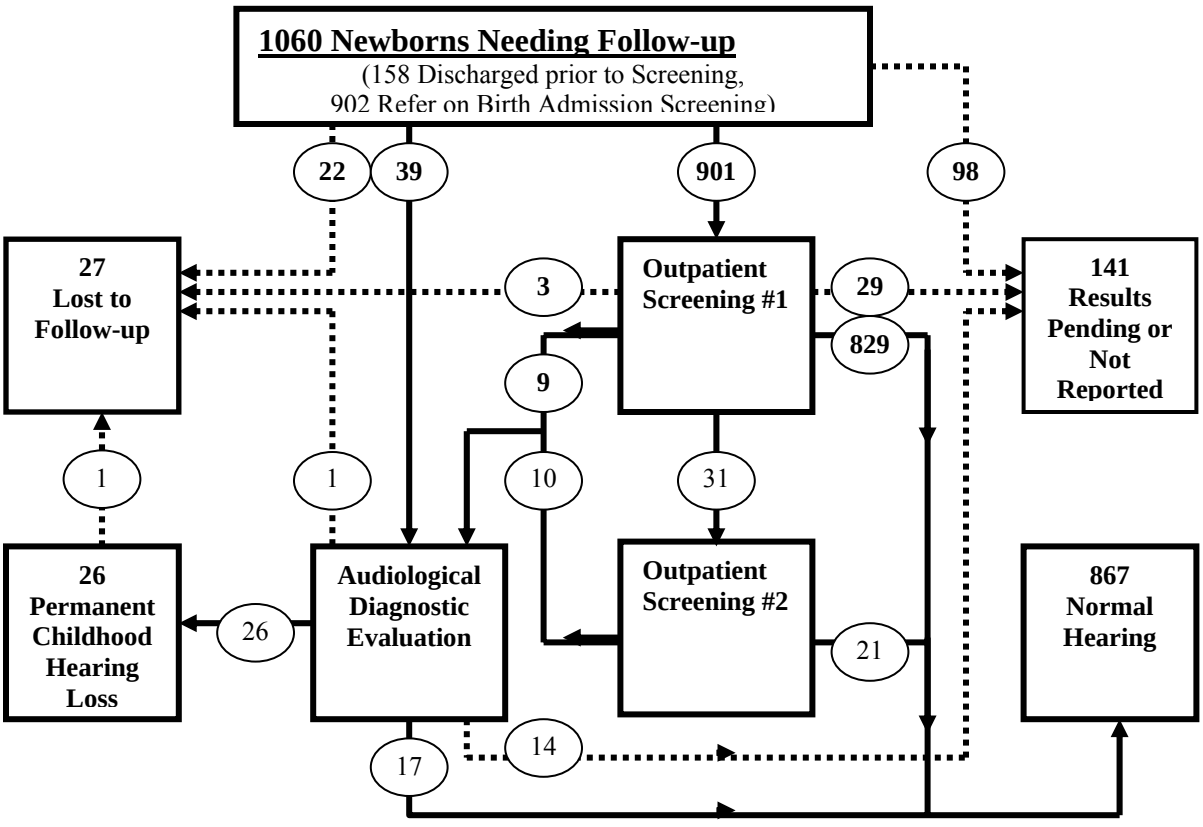


Diagram 1

Neb. Rev. Stat. §71-4742 states: "…it is the goal of this state to achieve a one-hundred-percent screening rate." While Nebraska has made great strides in developing a comprehensive newborn hearing screening system, there are also infants for whom the status of their hearing is not known. In 2004, there were 215 newborns whose hearing status has not been established:

- 19 of the 158 newborns who did not receive a hearing screening during birth admission also did not receive an initial hearing screening as an outpatient or the results were not submitted to NNHSP
- 79 of the 898 who "referred" on the hearing screening during birth admission either had no outpatient re-screening completed or the results were not submitted to NNHSP
- 29 who referred on the initial outpatient hearing screening either had no further follow-up evaluation completed or the results were not submitted to NNHSP
- 14 who were diagnosed with a hearing loss (12 with temporary conductive hearing loss due to middle ear dysfunction and 2 with unspecified type of loss) had no further follow-up re-evaluation completed or the results were not submitted to NNHSP
- 47 of the out-of-hospital births have not been screened or the results were not submitted to NNHSP
- 27 infants have been lost to follow-up (family moved, primary care physician for follow-up communication could not be identified, parental refusal of services)

Based on the analysis of the aggregate hospital reports and actual file counts, the hearing status of only 0.8% of the 26,443 newborns was not confirmed as either normal hearing or hearing loss present.

Timeliness of Follow-up Re-screening/Testing

To meet the state and national guidelines of "1-3-6" (hearing screening completed by 1 month, audiological diagnostic evaluation initiated by 3 months, early intervention initiated by 6 months), the timeliness of initiation of follow-up activities is an important aspect of the quality of services. In 2004, there were 805 newborns for whom follow-up re-screening and/or diagnostic evaluation commenced (this does not include those newborns discharged prior to screening). Of those, 597 (74.2%) had follow-up services initiated by one month of age. As Chart 3 displays, the peaks of follow-up activity occur at approximately one week, two weeks, and three weeks of age. The average age of follow-up service initiation for the entire group of 805 newborns was 30.6 days.

Of the total of 940 newborns for whom outpatient screenings and/or diagnostic evaluations were initiated, 135 of them had not received the newborn hearing screening during the birth admission and were discharged prior to screening. Of those, 77% (104) received the initial screening as an outpatient before one month of age. Although an initial outpatient newborn hearing screening by one month of age does not meet the intent of Neb. Rev. Stat. §71-4739 for each newborn to be screened during birth admission, it does meet the national Early Hearing Detection and Intervention goal of every newborn

having a hearing screening by one month of age.

Timeliness of Initiation of Follow-up Services

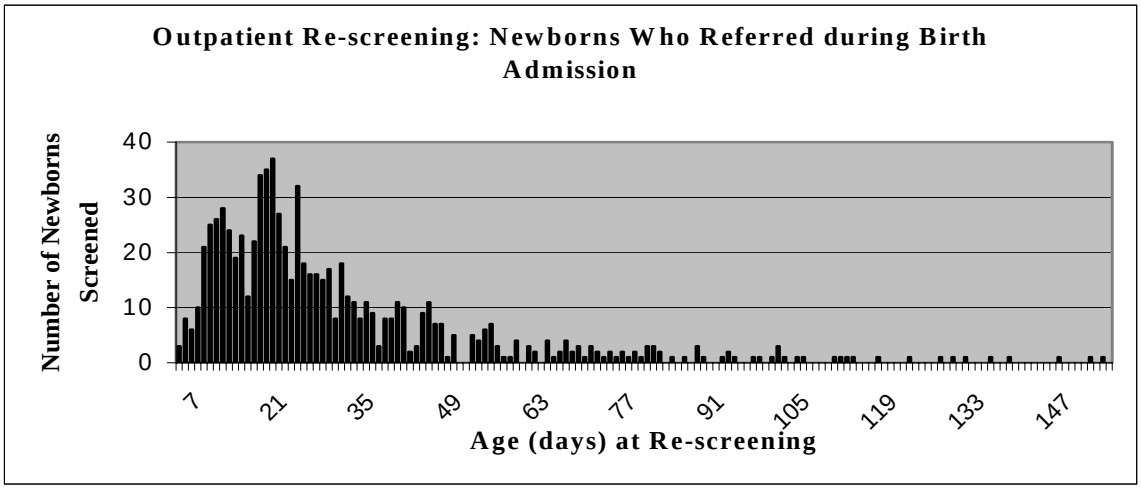


Chart 3

Diagnosis of Hearing Loss

The number of infants diagnosed with a hearing loss in Nebraska is reported in two ways: 1) aggregate reports submitted by audiologists of the number of infants shown to have a hearing loss based on follow-up tests (required by Neb. Rev. Stat. §71-4739) and 2) the individual diagnostic reports submitted to NNHSP by audiologists or primary care physicians. Statuatory authority to require audiologists to report on all newborns and infants that receive audiological evaluations does not exist, so a one-to-one correspondence between the individual results reported to NNHSP and the required annual aggregate reporting does not exist. Audiologists reported conducting 110 diagnostic evaluations of infants born in 2004, identifying 53 infants with hearing loss. The NNHSP received 58 patient-specific reports of the 110 evaluations and 46 patient-specific reports of the 53 infants identified with a hearing loss. Voluntary reporting of individual newborns and infants diagnosed with hearing loss has consistently increased from 39% in 2001 to 87% in 2004 (see Table 8).

Percentage of Patient-Specific Diagnostic Reports Received

	2001	2002	2003	2004
Percent of reports received for infants identified with hearing loss compared with aggregate reports	39%	76%	85%	87%

Table 8

Type and Degree of Hearing Loss

In 2004, based on individual reports submitted to NNHSP, there were 111 newborns who needed confirmatory testing beyond the initial outpatient re-screening to determine the

status of their hearing. The results of the 72 who referred on the first outpatient hearing screening were:

- 21 passed the re-screening
- 3 were lost to follow-up (moved, refused)
- 29 referred on the screening but either did not receive additional follow-up or the results were not reported to NNHSP
- 19 referred on the first or second outpatient screening and then received a diagnostic evaluation

Thirty nine (39) of the 111 newborns received a diagnostic evaluation as the initial step, bringing the total number of individual diagnostic reports to 58. The results of the 58 audiological diagnostic evaluations are:

- 17 had normal hearing established either initially or following medical management for middle ear dysfunction
- 14 are in follow-up for conductive hearing loss due to middle ear dysfunction or to complete the evaluation
- 1 was lost to follow-up (refused)
- 26 were diagnosed with a Permanent Childhood Hearing Loss

The targeted hearing loss for identification by newborn hearing screening programs is Permanent Childhood Hearing Loss (PCHL), a "permanent bilateral or unilateral, sensory or conductive hearing loss, averaging 30 to 40 dB or more in the frequency region important for speech recognition" (Joint Commission on Infant Hearing, 2000). The estimates of incidence of hearing loss in newborns range between 1 to 3 per thousand births nationally. Based on the birth rate in Nebraska during 2004 (26,443), an estimated 26 to 79 newborns would be identified with PCHL. Analysis of the patient-specific diagnostic reports submitted to NNHSP indicates that 26 infants born in 2004 have a PCHL. Analysis of the aggregate reports indicates that 29 infants meet the criteria for PCHL. Sixteen (59%) of the 26 infants identified with PCHL were identified prior to three months of age and the average age at identification was 98.4 days.

Type and Degree of Permanent Childhood Hearing Loss, 2004 (n=26)

Degree ► / Type ▼	Bilateral Mild–Moderate	Bilateral Severe–Profound	Unilateral Mild–Moderate	Unilateral Severe-Profound
Sensorineural	6	9	2	2
Conductive	0	-	4	-
Mixed	1	1	0	0
Auditory Neuropathy			1	

Table 9

Early Intervention

The purpose of the Infant Hearing Act (Neb. Rev. Stat. §71-4735) is to "obtain needed multidisciplinary evaluation, treatment, and intervention services at the earliest opportunity and to prevent or mitigate the developmental delays and academic failures associated with late detection of hearing loss." The Early Development Network,

Nebraska's Part C Early Intervention Program, has identified 17 (63%) of the 26 infants with PCHL to be eligible for Early Intervention services. Services were begun at an average of 157 days of age, meeting the benchmark of initiation of services by six months of age. Eleven of the infants had hearing impairment identified as the primary verified disability and the remaining six had a primary verification other than hearing impairment. Two of the infants identified with a PCHL were not eligible for services, the parents of one infant refused services, one family moved out-of-state, and six of the identified infants have not been evaluated.

Summary

- All the current birthing hospitals in Nebraska were conducting newborn hearing screening in 2004. All but two were conducting the hearing screenings during the birth admission.
- The benchmark of 95% of newborns having a hearing screening during birth admission by December 1, 2003 established by Neb. Rev. Stat. §71-4742 has been met. In 2004, birthing hospitals reported screening the hearing of 98.2% of newborns.
- The overall refer rate of 3.5% for initial hearing screening during birth admission was within national norms during 2004.
- The rate of reported follow-up re-screening and/or diagnostic evaluation has continued to improve, increasing from 63% in 2001 to 89% in 2004. This compares favorably with the national average of 55% for completion of the recommended follow-up testing.
- In 2004, follow-up re-screening occurred within one month of birth for 74% of those newborns for which follow-up activities were initiated. The average age at the time of the initiation of follow-up re-screening or diagnostic evaluation was 30.6 days.
- The percentage of patient-specific diagnostic evaluation reports submitted to NNHSP continued to increase from 39% in 2001 to 87% in 2004.
- The average age at diagnosis of hearing loss was 98.4 days for those reported to NNHSP in 2004 and 59% of the evaluations occurred within 3 months of birth.
- The incidence of Permanent Childhood Hearing Loss identified (1 per thousand in 2004) and reported to NNHSP appears to be within the anticipated range of 1 to 3 per thousand.

ACTIVITIES – 2004 to 2005

Funding

Funding from the Maternal and Child Health Bureau for the fourth year (2004-2005) of the Universal Newborn Hearing Screening (UNHS) project was awarded but there was a 2% reduction, which all UNHS programs received. The federal UNHS funds of $43,650 (plus $4,526 of carryover from 2003) comprised 58% of the NNHSP budget. The remaining 42% of the budget was $35,000 that was allocated from the Title V MCH Block Grant for the current fiscal year. Federal funds have decreased by approximately 65% since the first year of this project.

Application was made to the Maternal and Child Health Bureau on November 1, 2004, for an additional three years of funding. The grant application was fully funded for $125,000 per year, an increase of 250% over the previous year. The goals for the Universal Newborn Hearing Screening and Intervention grant are:
System Goal 1 – The hearing of all newborns in Nebraska will be screened during the birth admission or, if born out-of-hospital, by one month of age.
System Goal 2 – All newborns who "refer" on the initial outpatient hearing re-screening will complete an audiologic diagnostic evaluation prior to 3 months of age.
System Goal 3 – All infants with confirmed hearing loss will begin receiving early intervention services prior to six months of age.
System Goal 4 – All infants with a confirmed hearing loss will have a medical home.
System Goal 5 – Families of young children with a confirmed hearing loss will have access to a family-to-family support system.
System Goal 6 – The hearing of young children in Nebraska will be screened at various times prior to age 3.
System Goal 7 – Hearing health professionals will increase their capacity to provide appropriate services to young children.
System Goal 8 – NNHSP will provide an effective structure for the newborn hearing screening and intervention system in Nebraska.

Application was made to the Centers for Disease Control and Prevention on March 1, 2005, for an Early Hearing Detection and Intervention (EHDI) Tracking, Surveillance, and Integration cooperative agreement. The application will be funded at a reduced level of $145,850 per year for three years. The goals of the EHDI Tracking, Surveillance, and Integration cooperative agreement are:

Goal 1 – Hearing screening results will be electronically reported to NNHSP for all occurrent births in Nebraska.
Goal 2 – Pediatric audiologic evaluations, medical evaluations, and developmental outcomes will be electronically reported to NNHSP for young children identified with a hearing loss.
Goal 3 – The NNHSP data system, integrated with electronic birth certificate registry, will be electronically linked with related child data systems.
Goal 4 – A formative and summative evaluation of the NNHSP tracking, surveillance and integration project will be conducted and the results disseminated.

The carryover of funds from the Nebraska Health Care Cash Fund ($18,662) was approved to apply toward purchasing the integrated electronic data reporting and tracking system. Once again, Title V block grant funds of $1338 have also been committed toward the purchase of the system.

Electronic Data System

A commitment was made in April, 2004, to purchase the optional Newborn Hearing Screening Module from QS Technologies for integration with the new HHS Electronic Vital Records System. The integrated system will eliminate the need to manually record,

transmit, and track demographic information on each newborn who "refers" or is discharged without a hearing screening and will increase the accuracy, consistency, and timeliness of newborn hearing screening information provided to the NNHSP by birthing hospitals. Development of the module began in January, 2005, and the first demonstration was available for review in March. An initial review of the module included staff from a cross-section of birthing facilities and suggestions were incorporated into the first revision. Beta-testing is planned for the Summer, 2005, with full implementation by January, 2006.

Revision of Current Systems

As the electronic data system is developed, the NNHSP tracking and surveillance processes will be modified to take advantage of the features of the new system. Communication, reporting, and evaluation processes will be revised and enhanced. As part of the revision of reporting processes, particular attention will be paid to clarify the legislative language regarding reporting requirements to increase the accuracy and usefulness of the data.

Early Head Start ECHO Project

The National Center for Hearing Assessment and Measurement (NCHAM) selected Nebraska to participate in the next phase of their Hearing Head Start project. Funded by ACF/Head Start Bureau, five states have participated in previous phases; this phase expands to three additional states. The Early Childhood Hearing Outreach (ECHO) project trained a cadre of professionals in the hearing health and early childhood fields to train staff in Migrant, American Indian, and Early Head Start programs to screen the hearing of infants and toddlers using OAE screening equipment and to establish referral and follow-up protocols. The project began in September, 2004, with the training of a core group of trainers. ECHO training team members are:

- Dr. Donald Uzendoski, Boys Town Pediatric Primary Care, Omaha; Early Hearing Detection and Intervention Chapter Champion for Nebraska Chapter of American Academy of Pediatrics
- Darcia Dierking, Audiologist, Boys Town National Research Hospital, Omaha
- Nora Fuchs, Audiologist, Audio Logic PC, Columbus
- Charles Bee, Audiologist, Children's Hospital, Omaha
- Jennifer Rossi, Educator of the Deaf, The Omaha Hearing School, Omaha
- Martha Nash, Training Coordinator, Early Childhood Training Center, Omaha
- Jeff Hoffman, Program Manager/Audiologist, Newborn Hearing Screening Program

Thirty (30) Early Head Start staff members from Central Nebraska Community Services in Loup City and the Head Start Child and Family Development Program in Hastings have been trained to conduct OAE hearing screenings. Three additional Early Head Start grantees will be trained later in 2005. Screening equipment is being provided to the programs and over 600 infants and toddlers will be screened semi-annually.

Advisory Committee

The Advisory Committee of the Nebraska Newborn Hearing Screening Program has been re-activated and includes 16 of the original Advisory Committee members with six new members representing parents, family support, and education stakeholders. Five sub-committees have been formed to develop plans and protocols for NNHSP. The sub-committees are:

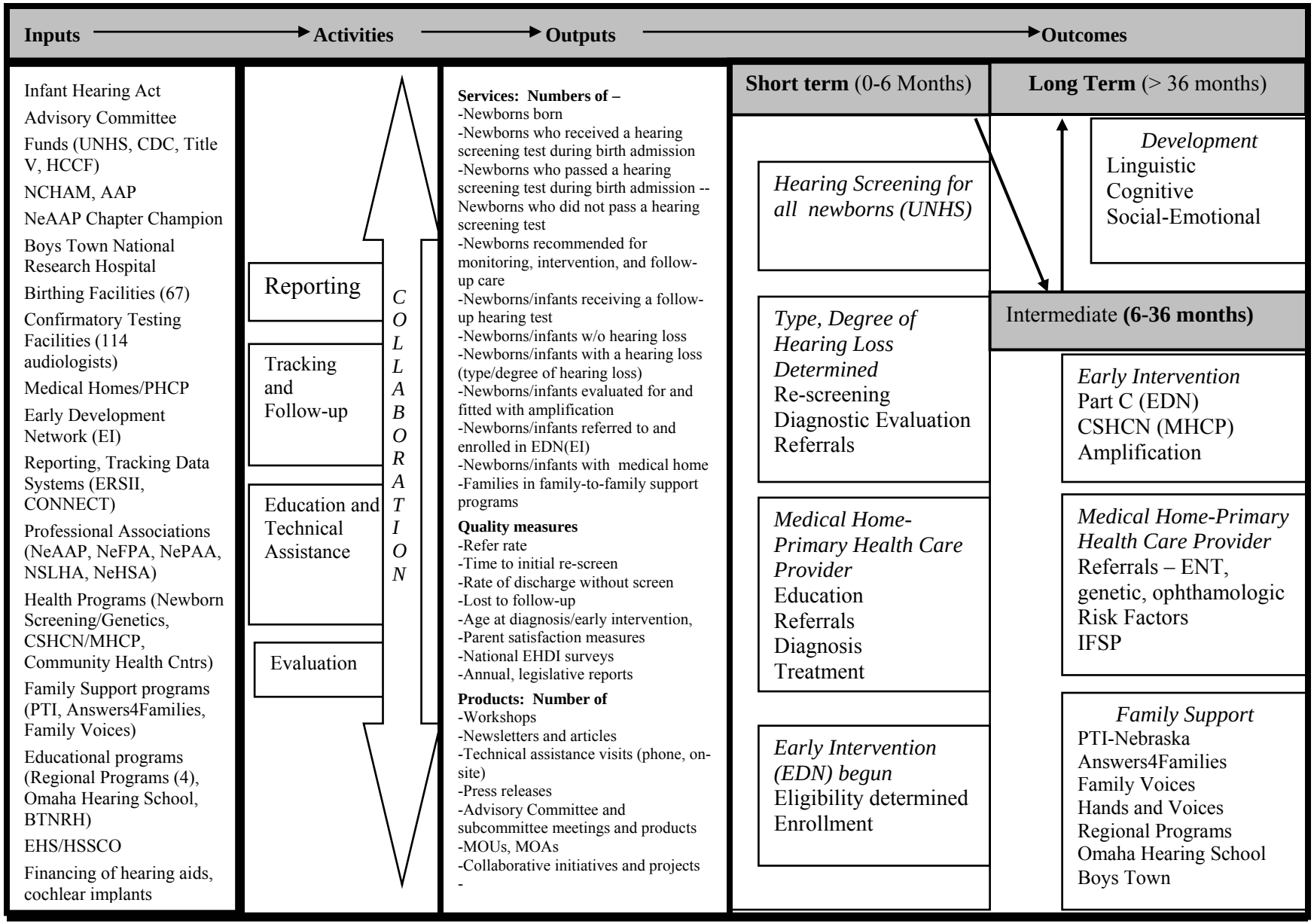
- Post-diagnosis referral and evaluation protocols for medical specialty evaluations
- Post-diagnosis referral and evaluation protocols for early intervention and audiological management
- Electronic data reporting system
- Review and revise current protocols for initial screening, re-screening, and audiological diagnostic evaluations
- Resources for parents with a child newly identified with a hearing loss

Some babies are born listeners . . .

Others need your help!

APPENDIX

Nebraska Newborn Hearing Screening Program – Logic Model



The staff of the **Nebraska Newborn Screening Program** are available to help with your questions at the numbers listed below:

Julie Miller, Newborn Screening/Genetics Program Manager (402) 471-6733
Krystal Baumert, Follow-up Coordinator (402) 471-0374
Kristen Strasheim, Follow-up Specialist (402) 471-6558
Mike Rooney, Administrative Assistant (402) 471-9731

Nebraska Newborn Screening Program
Department of Health and Human Services Regulation & Licensure
P.O. Box 95007
Lincoln, NE 68509-5007

The staff of the **Nebraska Newborn Hearing Screening Program** are available to help with your questions at these numbers listed below:

Jeffrey Hoffman, CCC-A, Newborn Hearing Screening Program Manager (402) 471-6770
Mike Rooney, Administrative Assistant (402) 471-9731
(See address above)

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